Ruptured internal carotid aneurysm resulting from neurofibromatosis: Treatment with intraluminal stent graft

Bertram L. Smith, MD, Carolyn E. Munschauer, BA, Norman Diamond, MD, and **Frank Rivera, MD,** *Dallas, Tex*

Purpose: **This report shows a method of treatment for life-threatening hemorrhage due to rupture of an aneurysm in the cervical internal carotid artery caused by neurofibromatosis.**
Methods: **Ten days after delivery of healthy twins, a 28-year-old woman with known neurofibromatosis had sudden massive swelling in the left neck. After initial tracheostomy, angiography confirmed rupture of the mid cervical internal carotid artery as well as contribution to the resultant pseudoaneurysm from external carotid branches. Treatment began with coil embolization of the external carotid branches. The internal carotid lesion, a defect approximately 1 cm in length, was then closed through use of two stent grafts, each made from Palmaz stents and 3-mm polytetrafluorethylene grafts predilated to 6 mm. The neck hematoma was then evacuated surgically.**
Results: **Completion angiography and computed tomographic scanning confirmed control of the hemorrhage. The patient survived neurologically intact with the exception of cranial nerve deficits caused by the hemorrhage. The tracheostomy tube was removed 3 weeks postoperatively. Follow-up computed tomographic scanning showed a gradual decrease in the size of the cervical soft tissue and no recurrent aneurysm.**
Conclusion: **Neurofibromatosis is a rare cause of aneurysmal degeneration of blood vessels. Repair of a ruptured cervical internal carotid artery aneurysm, though feasible, is difficult with stent grafts; however, this is a better option than surgical intervention in inaccessible vessels. (J Vasc Surg 2000;32:824-8.)**

Neurofibromatosis is an autosomal dominant disorder of variable penetrance that predominantly involves the peripheral nervous system with abnormal growth of neuroectodermal tumors. Arterial lesions have been variously described as involving small arterioles with concentric laminations of intimal proliferation and larger arteries with fibrous intimal thickening.[1-4] The most common clinical arterial lesion is renal artery stenosis.[5-7] Cerebrovascular abnormalities described include occlusion of major intracranial vessels, aneurysms, and arteriovenous malformations.[4,8-10] This report describes the repair of a ruptured cervical internal carotid aneurysm in a patient with known neurofibromatosis. Repair was accomplished with an endoluminal stent graft fashioned from a Palmaz stent covered with polytetrafluorethylene (PTFE). Because of the massive amount of hemorrhage and the location of the aneurysm, direct operative repair was potentially disastrous.

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Reprint requests: Bertram L. Smith, MD, 712 N Washington, Suite 509, Dallas, TX 75246.

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CASE REPORT

Ten days after the delivery of healthy twins, a 28-year-old woman with a history of neurofibromatosis and previous neurofibroma resected from the left posterior fossa at age 5 years had sudden massive swelling in the left neck. She had had a headache for the previous 2 days. Her family had noted a small mass in the left neck during her pregnancy but attributed no significance to it.

The patient was initially seen in an outlying emergency room where she had episodes of decerebrate posturing that were treated with diphenylhydantoin. She received an immediate tracheostomy at that hospital and was transferred to a second hospital for angiography. Angiography showed a ruptured aneurysm of the left internal carotid artery. She was transfused with blood products and transferred to Baylor University Medical Center by helicopter.

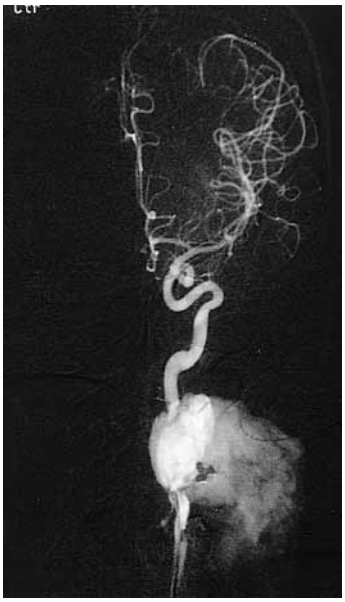

Fig 1. Left common carotid angiogram shows internal carotid extravasation.

Fig 2. Left internal maxillary artery selective catheterization reveals extravasation into aneurysm cavity.

Physical examination revealed an alert female patient with massive swelling of the left neck. The previously placed tracheostomy tube was deviated and near the level of the right mid clavicle. The tongue was protuberant and the eyes were swollen shut; however, her vital signs were stable. The blood pressure was 131/89 mm Hg. Pupils were equal in size, and the patient was neurologically intact.

She was taken immediately to the angiography suite. Angiography revealed a normal right carotid bifurcation without evidence of supply of the left carotid aneurysm from the right. Intracranially, the right anterior cerebral artery supplied the left anterior cerebral artery, but there was no supply from the right carotid to the left middle cerebral artery. Right and left vertebral angiography showed no contribution to the aneurysm. In addition, no collateral flow to the left middle cerebral circulation was identified from the posterior fossa.

Left carotid arteriography revealed an enormous multiloculated false aneurysm arising from the internal and external carotids (Fig 1). The extracranial left internal carotid artery had a large leak in its mid to distal portion, approximately 6 cm distal to the carotid bifurcation. Also noted was termination of the proximal internal maxillary artery in a portion of the aneurysm. The distal portion of the internal maxillary artery, distal to its connection to the pseudoaneurysm, was patent.

The internal maxillary artery leak was treated first. A selective catheter manipulated into the internal maxillary artery showed extravasation of contrast from a completely dehisced vessel (Fig 2). Many attempts were made to seek the distal internal maxillary artery distal to the origin of the extravasation. These attempts were not successful. A 5-mm Gianturco coil and then an 8-mm Gianturco coil were deployed into the internal maxillary artery with complete occlusion of the vessel at that time.

The distal internal carotid aneurysm rupture, which was the predominant source of the hemorrhage, was then treated. The internal carotid artery was selectively catheterized with a 5F vertebral catheter. The catheter was negotiated over a 0.035-in guidewire into the distal cervical portion of the vessel. At that point, a 0.035-in stiff Rosen wire was advanced through the catheter and the catheter was removed.

A stent graft was fashioned as follows: a segment of uncoated, thin-walled 3-mm PTFE graft was predilated to 6 mm with a balloon dilatation catheter. A Palmaz 394 stent was partially expanded. The PTFE graft was then sewn to the stent at the leading edge and the trailing edges with a 6-0 polypropylene suture on a noncutting needle. Two sutures were placed at the leading edge and two were placed at the trailing edge.

A 6-mm × 4-cm balloon dilatation catheter was then backloaded through a 10F guiding catheter. The stent graft was crimped on the balloon portion of the catheter and withdrawn into the guiding catheter. Heparin (3000 units) was given intravenously. A 12F sheath had been placed in the left groin over the Rosen wire. Through the sheath, the 10F guiding catheter with a stent graft apparatus was advanced over the guidewire to the distal left internal carotid artery. Arteriography was then performed to position the stent graft. It was then inflated to 6 mm.

After stent graft placement, arteriography showed that there was still supply to the aneurysm in the distal cervical portion of the internal carotid. A second stent graft was fashioned in the same way as the first; however, the stent used was a Palmaz 204. After the PTFE materi-

Fig 3. Completion angiogram shows successful deployment of second stent in internal carotid.

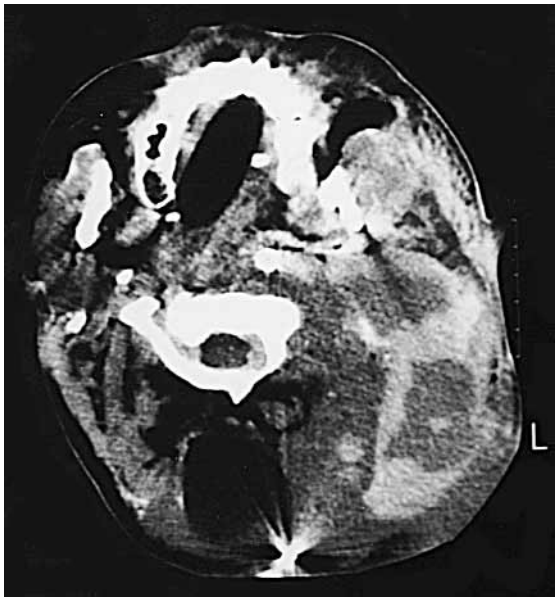

Fig 4. Head CT shows internal carotid stent and large paracervical hematoma (note contrast in hematoma).

al was sewn to the stent, it was loaded onto a 5F 6-mm × 2-cm balloon catheter. The guiding catheter was advanced over the guidewire, and the stent graft was deployed in an overlapping fashion over the initial stent graft onto the distal cervical portion of the internal carotid artery. The stent grafts were redilated to 6 mm throughout their course. Arteriography showed excellent control of the pseudoaneurysm in the neck (Fig 3) but late retrograde filling of the left internal maxillary artery, which emptied into the aneurysm.

At that point, a 5F vertebral catheter was manipulated through the previously placed internal maxillary artery coil. Attempts were made to seek the distal internal maxillary artery, but the catheter and wire persistently ended up in the aneurysm. Further attempts were unsuccessful.

A 5F catheter was then directed into the lingual artery. An attempt was made with a microcatheter and guidewire to enter the internal maxillary artery retrograde. The microguidewire actually did traverse a lingual artery branch, ascend superiorly, and enter the internal maxillary artery. However, the microcatheter would never follow the guidewire.

Next, a 6F sheath was placed in the opposite right femoral artery. A 5F catheter was advanced to the internal maxillary artery proximally and then into the pseudoaneurysm. That catheter was exchanged over a long guidewire for a 6F guiding catheter with a gooseneck snare having a diameter of 25 mm. Attempts were made to open the gooseneck snare and ensnare the micro-guidewire placed from the left femoral puncture into the distal maxillary artery. These attempts were unsuccessful. The patient was taken from the vascular and interventional radiology section for computerized tomography (CT) scanning.

CT showed a large, amorphous soft tissue mass extending from the skull base inferiorly to a point approximately 1.5 cm below the hyoid bone. This lesion measured 10 cm in maximum diameter, 7.5 cm in the craniocaudal dimension, and 8 cm in the anteroposterior dimension (Fig 4). The lesion enveloped the lateral and posterior walls of the left internal carotid artery, where a stent graft was in place. The mass infiltrated the pinna of the ear and diffusely involved the posterior left masticator space and posterior triangle and virtually replaced the upper anterior triangle of the neck.

The patient had also had a previous neurofibroma resected from the posterior fossa, and the occipital bone was dehisced in this area—a chronic change. There was no evidence of intracranial malignancy or CT findings to suggest neurofibrosarcoma of the left neck.

The patient was then taken to the operating room to decompress and evacuate the large residual hematoma and remove some of the pressure from the contained hemorrhage in the neck. At the time of surgery the tissue of the neck was friable and suture ligation of the internal maxillary artery bleeder was performed. Because of tissue friability, exposure of the distal internal carotid was not possible despite stent graft control of the ruptured aneurysm. Because of the friable tissue present, only fine Prolene (Ethicon, Inc, Somerville, NJ) could be used for suture ligation. Use of the argon-beam coagulator, meticulous technique, and minimal manipulation of the tissues resulted in an intraoperative blood loss of 2000 mL. The wound was closed over a drain.

The postoperative course was relatively uncomplicated. The patient remained on mechanical ventilator support for 48 hours. She remained alert, moving all extremities with-

out deficit. Tube feedings were begun for nutrition. The speech pathology department was consulted for assistance with the patient's swallowing and communication. She was transferred out of intensive care on postoperative day 4. A pathologic specimen from the operative site showed neurofibroma with no evidence of malignancy.

On postoperative day 16, the patient's tracheostomy tube was exchanged for a smaller tube, and she began tolerating a soft diet by mouth. She was discharged on postoperative day 18.

Follow-up CT scans have shown gradual resolution of the mass effect in the left neck, and the patient has had no further episodes of bleeding 18 months after the original event (Fig 5). Follow-up duplex scans at 6-month intervals have shown a patent left internal carotid artery without abnormal flow velocities.

Fig 5. Follow-up CT scan 3 months after hospital discharge.

DISCUSSION

Neurofibromatosis I is an autosomal dominantly inherited disorder characterized mainly by hamartomatous and neoplastic proliferations of cells of the peripheral and central nervous system in a broad range of clinical patterns involving the skin and eye, brain and spinal cord, and, minimally, the long bones and arteries.[1] The arterial involvement may be with stenotic lesions or aneurysmal ones and has been variously studied, but because of the lack of arterial lesions and patients involved, a systematic, widely accepted categorization does not exist.

In 1945, Reubi[3] characterized three types of arterial lesions. These arterial abnormalities occurred in vessels less than 1 mm in diameter and predominantly involved the kidney. Reubi described (1) intimal proliferation leading to occlusion of the vascular lumen, (2) fibrous thickening of intima with fragmentation of the muscularis and elastica, causing aneurysmal dilatation of the vessel wall, and (3) fusocellular nodularity compromising the strength and integrity of the vessel wall.

In 1973, Salyer and Salyer[2] hypothesized that a proliferation of Schwann cells within the arterial wall was the cause of all of the vascular abnormalities observed. They believed that the different types of vascular abnormalities represented the primary proliferation of Schwann cells, a secondary degenerative change, or the healing process. In another review (in 1974), Greene et al[4] suggested that there were two primary types of vascular lesions associated with neurofibromatosis. The first type affected large vessels with perivascular neurofibromas or ganglioneuromas associated with degenerative changes in the adjacent vessel wall; the second type was present in small vessels and consisted of smooth nodular aggregates of smooth muscle cells that Greene et al believed represented mesodermal dysplasia.

The most common clinical presentation for vascular involvement by neurofibromatosis is with renal artery stenosis and secondary hypertension.[5-7,11] Cerebrovascular abnormalities include occlusion of major intracranial vessels, arteriovenous malformations, and intracranial aneurysms.[4,8-10] Previous reports of carotid artery aneurysms are rare—saccular aneurysms in the common carotid artery and an asymptomatic aneurysm of the petrous carotid artery.[8-10] The literature is not helpful in assessing the natural history of cerebrovascular lesions, largely because of their small number.

In the current article, the patient had a mass in the left neck that had been noticed by her family during her pregnancy. This probably represented a saccular aneurysm of the internal carotid artery. Because of the location of this aneurysm and the inability to attain distal arterial control with operative exposure, direct operative repair would have been treacherous. Moreover, the friability of the tissues noted at the time of hematoma evacuation made simple manipulation of the vessels impossible without significant blood loss. Ligation of the internal carotid artery or intraluminal balloon occlusion might have been possible. The patient did not have angiographic evidence that her left hemisphere was perfused with absence of flow in the left internal carotid. Internal carotid stump

pressures were not calculated. Moreover, because the contribution of the external carotid branches to the pseudoaneurysm and collateral flow from the right external carotid made mere internal carotid ablation unattractive and potentially not therapeutic, a trial of balloon occlusion of the internal carotid was not done.

Success of therapy in this particular case depended on the feasibility of placement of the second intraluminal stent graft when the first stent graft did not completely obliterate the hemorrhage from the internal carotid. Reports of covered stent graft placement in the internal carotid artery are few and reveal the risk of both embolization and graft occlusion.[12,13] The patient's cranial nerve deficits, glossopharyngeal and hypoglossal, resolved completely, with the exception of the left hypoglossal. Eighteen months after the procedure she swallows without difficulty but continues to have deviation of the tongue to the left. Follow-up physical examination, duplex scanning, and CT scans have shown no evidence of recurrent aneurysm, no arterial venous malformation or neurofibroma in the neck, and no evidence of malignancy. Long-term antiplatelet therapy was chosen for this patient; the results have been reasonable thus far.

In conclusion, arterial manifestations of neurofibromatosis I are unusual. Aneurysmal arterial degeneration is more unusual. Rupture of aneurysms in patients involved with neurofibromatosis is rare. In this instance, rupture of the internal carotid artery in a patient with neurofibromatosis was a potentially lethal event corrected only with successful placement of an intraluminal stent graft in the internal carotid. Although the immediate results were dramatic and early postoperative follow-up has been unremarkable, long-term results remain to be evaluated.

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